Newborn Screening Quality Assurance Program

PROFICIENCY TESTING — **Sickle Cell Disease and Other Hemoglobinopathies**

Volume 15, No. 4 | Quarter 4 | November 2005

INTRODUCTION

On October 3, 2005, we distributed to all active participants the Quarter 4 proficiency testing (PT) panel consisting of five dried-blood-spot (DBS) specimens for sickle cell disease and other hemoglobinopathies. A total of 70 PT panels were mailed by overnight FedEx mail. The packages went to 54 domestic laboratories and 16 foreign laboratories. The specimen panel consisted of five DBS specimens prepared from umbilical cord blood. This PT report is a compilation of all data reports for hemoglobinopathy testing received from participants by the designated deadline date. We distribute this quarterly report to all participants, state laboratory directors, and to program colleagues by request. We received data reports from 61 newborn screening laboratories. There were 9 laboratories that did not report this quarter. We requested that participants assay all survey specimens by the analytic schemes they routinely use and report for each specimen the presumptive phenotype, the presumptive clinical assessment, and any other clinical classifications that they deem consistent with their analytic results and program operations.❖

PARTICIPANTS' RESULTS

The certification report listing hemoglobins (Hbs) by phenotype and their presumptive clinical assessments appears on page 2.

The frequency distribution of reported phenotypes and presumptive clinical assessments appears on page 3.

The individual data verification for each laboratory with evaluation comments appears on page 4.❖

The NSQAP will ship next quarter's PT specimens on January 9, 2006.❖

SPOTLIGHT

Meetings

The 29th Annual Meeting of the National Sickle Cell Disease Program will be held in Memphis, TN, at the Peabody Hotel from April 8 – 12, 2006. This year the program will be hosted by the Comprehensive Sickle Cell Center at St. Jude Children's Research Hospital. For more information visit us at http://www.stjude.org/sickle-cellconf.

Awards

Congratulations to William (Jerry) Callan, Ph.D., Director of the Bureau of Clinical Laboratories, Alabama Department of Public Health. The 53rd Annual CDC & ATSDR honor awards ceremony held September 28, 2005, recognized Dr. Callan for his exceptional dedication toward ensuring the availability of evaluation materials for public health programs worldwide to monitor the effectiveness of sickle cell disease screening methods. Dr. Callan and his wife, Sharon, attended the ceremony in Atlanta, Georgia; where he was presented with the Partners in Public Health Improvement Award.❖

Dr. Jerry and Sharon Callan

ACKNOWLEDGMENTS

The specimens for this survey were prepared from umbilical cord blood samples supplied by Alabama State Public Health Laboratory.❖

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CDC
CENTERS FOR DISEASE CONTROL AND PREVENTION

Newborn Screening Quality Assurance Program
Sickle Cell Disease and Other Hemoglobinopathies

Specimen and Lab Certification

Year: 2005 Quarter: 4

Presumptive Clinical Phenotypes

	Specimen 4531	Specimen 4532	Specimen 4533	Specimen 4534	Specimen 4535
Expected Presumptive Phenotype	FAC	FSC	FA	FS	FAS
Accepted Presumptive Phenotypes	FCA		FAV FA + Fast		FSA

Presumptive Clinical Assessments

	Specimen 4531	Specimen 4532	Specimen 4533	Specimen 4534	Specimen 4535
Expected Presumptive Clinical Assessment	03	05	01	04	02
Accepted Presumptive Clinical Assessments	15		22,20, or 19	13*	13*

* Screening methods may not distinguish between homozygous S and Hb S + beta^0 thalassemia unless PCR is used.

01 Normal--no abnormal Hb found
02 Hemoglobin S carrier
03 Hemoglobin C carrier
04 Hemoglobin S, S disease (Sickle cell anemia)
05 Hemoglobin S, C disease
06 Hemoglobin S, D disease
07 Hemoglobin S, O disease
08 Hemoglobin D carrier
09 Hemoglobin E carrier
10 Hemoglobin G carrier
11 Hemoglobin O carrier
12 Hemoglobin S, E disease
13 Hemoglobin S Beta-thalassemia
14 Hemoglobin E Beta-thalassemia
15 Hemoglobin C Beta-thalassemia
16 Alpha-thalassemia (Bart's Hb)
17 Transfused infant
18 Hemoglobin E, E disease
19 Combination one or more
20 Assessment is not listed
21 Unsatisfactory specimen
22 Unidentified Variant
23 Hemoglobin E Alpha-thalassemia
24 Hemoglobin D Beta-thalassemia
(NE) Specimen not evaluated

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Frequency Distributions

Year: 2005 Quarter: 4

Phenotypes

Specimen Number	Hemoglobin Phenotypes	Frequency Distributions
4531	AFC	1
	FAC	59
	FCA	1
4532	FAS	1
	FCS	6
	FSC	54
4533	FA	48
	FA+Fast	2
	FAJ	3
	FAU	1
	FAV	6
	FAX	1
4534	FS	61
4535	AFS	1
	FA	1
	FAS	59

Clinical Assessments

Specimen Number	Presumptive Assessments	Frequency Distributions
4531	02 Hemoglobin S carrier	1
	03 Hemoglobin C carrier	59
4532	02 Hemoglobin S carrier	2
	05 Hemoglobin S, C disease	58
4533	01 Normal	48
	19 Combination one or more	1
	20 Assessment is not listed	2
	22 Unidentified Variant	9
4534	02 Hemoglobin S carrier	1
	04 Hemoglobin S, S disease (Sickle cell anemia)	59
4535	01 Normal	1
	02 Hemoglobin S carrier	59